UROONCOLOGY

Case Report

Renal cell carcinoma in ectopic-pelvic kidney: A rare case with review of literature

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ABSTRACT

The incidence of renal cell carcinoma in a pelvic kidney is rare, and has only been reported in a small number of cases. We report a 50-year-old male patient presented with vague abdominal pain and without hematuria. CT scan showed large heterogeneous soft tissue mass arising from a left pelvic kidney with areas of necrosis measuring 8.4x8.4x7.5 cm. Histopathology after radical nephro-ureterectomy showed grade II clear-cell renal carcinoma. Renal cell carcinoma of ectopic kidney is a rare disease. Even though the presentation might be atypical and challenging, the treatment strategy is still the same as for tumors of orthotopic kidneys.

Keywords: Ectopic kidney; pelvic ectopia; pelvic kidney; renal cell carcinoma; renal mass.

Introduction

Renal Cell Carcinoma (RCC) is the most common solid tumor affecting adults, accounting 2% of the worldwide and in Europe.[1] The diagnosis of RCC can be challenging as only almost 10% of the patients present with the classic triad of flank mass, flank pain and hematuria and thus it may preset with diverse manifestations including weight loss, general weakness, malaise, hypertension and hypercalcemia. Renal tumors of ectopic kidney are extremely rare. We report a case of a left pelvic kidney with a large renal cell carcinoma diagnosed in a patient presented with abdominal pain but without hematuria.

Case presentation

A 50-year-old male smoker known to have hypertension well controlled with valsartan/hydrochlorothiazide (80/12.5 mg) presented to the outpatient clinic in June, 2016 complaining of vague abdominal pain of two weeks duration. He described a discomfort in the lower abdomen which was not associated with nausea, vomiting or hematuria. General examination showed a male patient with BMI of 23.4 kg/m^2 (weight: 80 kg, height 1.85 m) and abdominal examination revealed a palpable mass in the lower abdomen. Routine laboratory workup, including complete blood count, renal, and liver function test results, and electrolytes were within normal range and urinalysis showed RBCs of 1-2 per HPF. Ultrasonography of the abdomen and pelvis showed left pelvic kidney with a large renal mass. Contrast-enhanced CT scan of abdomen and pelvis showed large heterogeneous soft tissue mass arising from the left pelvic kidney with areas of necrosis measuring 8.4x8.4x7.5 cm demonstrating significant corticomedullary phase enhancement with no metastasis. (Figures 1, 2). The vascular supply was provided from a single left renal artery arising from the left common iliac and a single left renal vein draining into the left common iliac vein.

After a multidisciplinary team discussion and the patient's informed consent, the patient underwent radical left pelvic nephro-ureterectomy via transperitoneal approach in July, 2016. The postoperative period was unremarkable. Specimen showed a unifocal tumor limited to the kidney and histopathology revealed clear-cell renal carcinoma grade II with negative margins. TNM staging was pT2 Nx M0 (Figure 3). The patient went abroad and he was instructed to attend his follow-up visits in the country of his residence.

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Table 1. Review of cases from the literature

Reference, year	Age, Gender	Presentation	Comorbidities	Side	Vascularity	Surgery	Histopathology	Metastasis	Follow up
Fischer et al.[3], 1999	48, Male	Hematuria	None	Right	Single renal artery originating from the common iliac artery. Single renal vein draining into the right external iliac vein	Open radical nephrectomy	RCC, Clear, Fur 1/4	Tumor thrombus in the renal vein	Not reported
Terrone et al.[4], 2004	52, Male	Abdominal pain	Not reported	Left	NA	Open radical nephroureterectomy	RCC, Fur 4/4	No	9 months after surgery the patient had no recurrence
Karaman et al.[5], 2009	66, Male	Weight loss, abdominal pain and palpable mass	NA	Left	Renal artery originating from left common artery	Nephrectomy	RCC, Fur 3/4	No	The patient is under control program without any evidence of local recurrence or metastasis for 24 months.
Mahmoudnejad et al.[6], 2009	53, Female	Abdominal pain	Not reported	Left	Two renal arteries originating from left internal iliac artery. Single vein identified	Open radical nephrectomy	RCC, Clear, Fur 2/4	No	Within postoperative first year the patient underwent physical examinations and metastatic work up and no delayed complications occurred.
Dash et al.[7], 2010	55, Male	Acute urine retention and weight loss	Posterior cortical atrophy	Left	NA	No surgical intervention was done	RCC (post mortem)	No	Patient did not have surgery and died four months after diagnosis
Baldie et al.[8], 2012	61, Male	Hematuria and irritative symptoms	Morbid Obesity	Left	Two renal arteries originating from left common iliac artery. Vein drains into left internal iliac	Open radical nephroureterectomy	RCC, Papillary, Fur 3/4	No	Patient is on dialysis and recurrence-free for 4 years since surgery
Nowroozi et al.[9], 2015	53, Female	Hematuria	None	Left	NA	Open nephrectomy	RCC, Clear, Fur 4/4	No	No local recurrence was detected for 18months after surgery
Parashari et al.[10], 2015	45, Male	cervical spondylitis myopathy (cervical metastasis)	Smoking	Left	Arterial supply from the left common iliac artery	No surgical intervention was done	NA	Vertebra	Not reported

Table 1. Review of cases from the literature (continued)

Reference, year	Age, Gender	Presentation	Comorbidities	Side	Vascularity	Surgery	Histopathology	Metastasis	Follow up
Tanwar et al.[11], 2016	61, Male	Gross hematuria	Not reported	Right	Single renal artery. Two veins draining into the right internal iliac and left common iliac	Radical nephroureterectomy with thrombus extraction	RCC, Clear with sacromatoid differentiation, Fur 3/4	Tumor thrombus invading both iliac veins	The patient is on surveillance for renal carcinoma and is following the protocol strictly.
Present case, 2016	50, Male	Abdominal pain	Smoking	Left	Single renal artery origination from the left common iliac artery. Single vein draining into the left common iliac	Radical nephroureterectomy	RCC, Clear, Fur 2/4	No	The patient went outside the country and is to be followed up in the country of his residence.

NA: not available

Figure 1. Contrast-enhanced abdominopelvic CT, coronal view showing a large heterogeneous soft tissue mass with dimensions of 8.4x8.4x7.5 cm arising from left pelvic kidney

Figure 2. Contrast-enhanced abdominopelvic CT, Axial view showing a large heterogeneous mass with central necrosis arising from left pelvic kidney

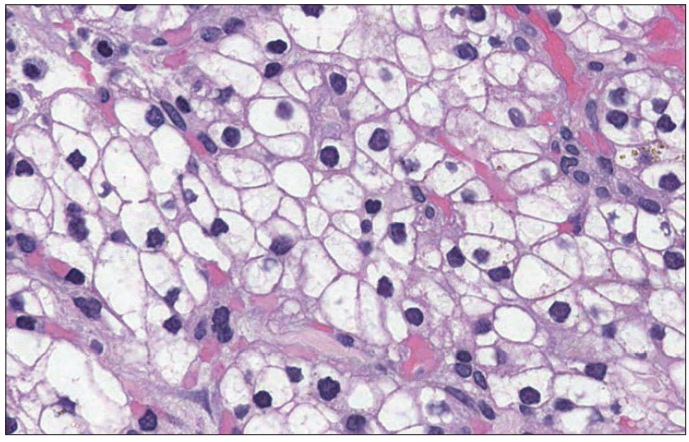

Figure 3. Clear-cell renal cell carcinoma

Discussion

Ectopic pelvic kidney is a rare developmental anomaly. During embryological life metanephros from which the kidney originates is located in the sacral region. During development, both kidneys ascend to their usual location in the retroperitoneal area. Early arrest or failure of ascent results in ectopic kidney.[2]

An ectopic kidney is mostly discovered incidentally during investigation for other reasons. However, it may be associated with other anomalies involving other systems including skeletal, urinary and cardiovascular systems.[2]

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The average incidence of pelvic kidney ranges between 1/2200 and 1/3000. The relationship between malignancy and ectopic kidneys is still unclear. To our knowledge, three cases of urothelial carcinoma and nine cases of renal cell carcinoma (RCC) in ectopic pelvic kidneys have been reported in the literature and there does not appear to be an increased risk of malignancy in ectopic kidneys. These cases are summarized in Table 1.[3-11]

The rarity of such cases in daily clinical practice made it a challenge for the urologists. Most cases of ectopic kidneys present with atypical presentations. However, the existence and availability of modern radiological imaging techniques increased the detection rates of such anomalies over the years.
The only reported masses in pelvic kidneys are renal cell carcinomas and upper tract urothelial carcinomas, so the suggested treatment options for tumors in pelvic kidneys are the same as tumors in orthotopic kidneys. The treatment modality and surgical approaches depend on the site, the clinical stage of malignancy, comorbid conditions and the surgeon's experience. Radical nephro-ureterectomy appears to be the standard treatment for localized tumors in ectopic kidneys.

The vascular supply of pelvic kidneys is complex and it is provided from the adjacent blood vessels. So the surgical approach to ectopic kidneys merits caution because of uncertain vascular anatomy.[6] It is mandatory to perform a detailed preoperative vascular evaluation, and also careful exploration during surgery in order to avoid vascular injury. Follow-up is required to detect any metachronous renal tumors, local recurrence and distant metastases.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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